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# SEVERE INTELLECTUAL DISABILITY IN DANDY WALKER VARIANT: A CASE REPORT

Psychiatry

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KEYWORDS

## INTRODUCTION :

Dandy walker syndrome was coined by **Walter Dandy and Arthur Walker** (1990 ). It is a rare congenital malformation of cerebellum. Diagnostic criteria according to Dandy and Blackfan's triad includes cystic dilation of the fourth ventricle, partial or complete absence of cerebellar vermis, hydrocephalus. Dandy walker syndrome is a genetically sporadic disorder that occurs in 1/35000 live births mostly in females. Dandy walkar variant is a milder form of Dandy Walker Syndrome with variable hypoplasia of cerebellar vermis, with or without enlargement of cisterna magna (communication between the 4th ventricle and the arachnoid space).No hydrocephalus. Several case reports have shown Dandy Walker variant malformation associated with psychotic symptoms, mood symptoms, obsessive compulsive symptoms, hyperactivity and impulsive behaviour. Intellectual disability with IQ ranging from 25-79 is also reported in few cases with Dandy Walker variant.

### Case Presentation:

### History:

A 9 year old girl was brought by her father to psychiatry OPD of SVRRGGH for disability certification, with complaints of hyperactivity, poor attention, emotional disturbances, walking difficulties. She was born out of consanguinous marriage, FTND, NBW, she had speech delay, poor scholastic performance, with no psychiatric illness in family. No history of fever, convulsions, hypotonia.

### On examination:

microcephaly, clinodactyly,strabismus,growth retardtion, bilateral club foot .Normal tone and power in all the limbs.

### Investigations:

All routine blood investigations – CBC, LFT, RFT, S.Electrolytes, viral screening where within normal limits. MRI BRAIN –Pontine hypoplasia, diffusely thin corpus callosum, posterior periventricular hyperintensities, mild lateral ventricular and sulcal prominence.CT BRAIN PLAIN –Hypoplastic cerebellar vermis .

### Treatment given:

- Tab.Risperidone 1mg OD
- Tab Atomoxetine 18mg OD were started for hyperactivity and behavioural problem

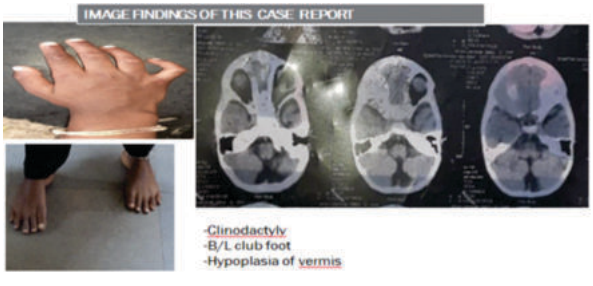


**Figure 1**

## DISCUSSION :

Multiple congenital defects may shorten life span ,longetivity depends on severity of syndrome and associated malformation. Some cases never achieve normal intellectual disability. Upto half of the affected individuals have intellectual disability that ranges from mild to severe and those with normal intelligence may have learning disabilities.Majority of individuals with DWS malformations or signs and symptoms caused by abnormal brain development are present at birth or present within the first year of life.Problems related to hydrocephalus or complications of treatment are the most common cause of death in DWS.

## CONCLUSION :

Understanding the clinical presentation of Dandy walker variant , evaluating the IQ and understanding the behavioural symptomology underlying this disease helps in improving the efficiency of treatment. Intellectual disability is managed by developing a comprehensive management plan ,which require input from multiple disciplines, including community services that provides social support care and respite care for families affected by intellectual disability.

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